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Case Reports

Arteriovenous malformation of the hand: challenges in diagnosis and management

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Abstract
Vascular malformations are not often considered in the differential diagnosis of digital lesions presenting acutely, but they do occur, and can present difficulties in diagnosis leading to delays in definitive treatment. We highlight the challenges in managing such patients, who often require multi-disciplinary and multi-centre care, and we hope to increase awareness of this condition.

Case History
A 36 year-year-old, right handed, unemployed man presented to his General Practitioner with what appeared to be a small abscess at the distal tip of his left thumb, a month after a trip to Goa. There was a vague and unconvincing history of possible minor trauma whilst he was abroad, and there was no other history of note. In the absence of a response to a course of oral antibiotics, he was admitted to our hospital on at least five separate occasions over a period of weeks under the care of a number of different surgical teams. In the absence of any response to intravenous antibiotics and surgical debridement during this period, he was subsequently referred to our hand (plastic surgery) unit. At this time, his thumb was painful and swollen with discoloration extending to the thenar eminence, a necrotic tip, and surgical scarring (figure 1).

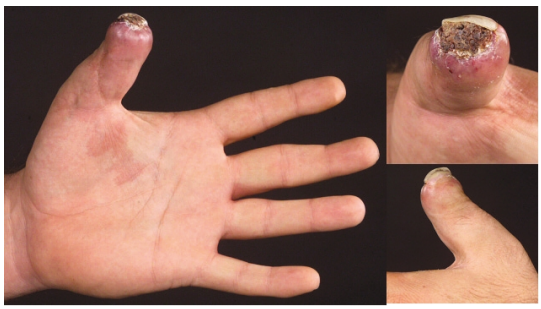

Figure 1. *Multi-angle photographs of the left hand and thumb at time of presentation. Note the discolouration over the thenar eminence.*

Systemic examination and his medical history were unremarkable, although he was a smoker, but routine investigation showed a mildly elevated C-Reactive Protein (15 mg/l) and leucocytosis (13.1 x 109/l). Thrombophilia screen and immune profiles were negative. A plain radiograph (figure 2) showed areas of lucency within the distal and proximal phalanges.

Figure 2. *A plain radiograph of the thumb shows diffuse soft tissue swelling of the thumb. A tubular lucency is seen in the proximal phalanx suggestive of a vascular impression. The presence of soft tissue swelling inassociation with vascular markings raises the possible diagnosis of an arteriovenous malformation.*

On the basis of these findings, treatment was undertaken with further surgical debridement with specimens sent for culture and histology and subsequent broad-spectrum intravenous antibiotics. Microbiological cultures yielded no growth at any time (including atypical organisms and tubercle bacilli), but histopathology reported some tissue containing richly vascular fibrous areas.

Initial management proved unsuccessful, and there was little clinical improvement. At this stage, differential diagnoses that had been considered included:

- Osteomyelitis (bone lucency with osteopoenia and raised inflammatory markers)
- Buerger's disease (a smoker with digital gangrene)
- Ischaemia from emboli (well demarcated ischaemic-looking digital lesion)
- Tropical disease, such as tuberculous dactylitis[1] (recent travel to Goa)

Detailed vascular clinical examination was normal, as were echocardiography and electrocardiography, and an MRI scan was subsequently requested. This revealed a large vascular malformation of the thumb with a significant intraosseous component (figure 3), features which explained the lucent areas of the initial plain radiograph (figure 2).

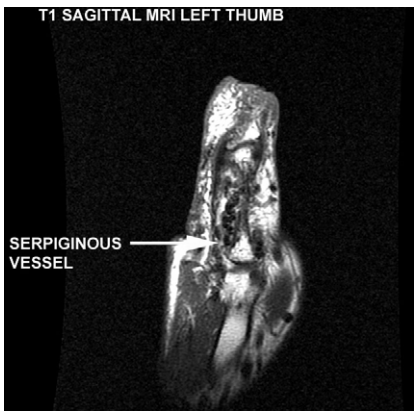

Figure 3. *MRI confirmed a large vascular malformation of the left thumb bydemonstrating large serpiginous vessels in both the soft tissues and the proximal and distal phalanges. The largest intraosseous vessel corresponds with the lucency shown on the plain film (figure 2).*

Coronal studies of the whole hand showed some abnormal vessels of the index, middle, and ring fingers also. The patient was subsequently referred to the Hammersmith Hospital for angiography with a view to therapeutic embolisation. The angiographic images are striking (figure 4), and limited embolisation was undertaken at the same procedure by puncturing some of the arteriovenous communications within the proximal phalanx via a percutaneous transosseous approach and occluding them with sodium tetradecyl sulphate (figure 5) resulting in significant clinical improvement over subsequent weeks.

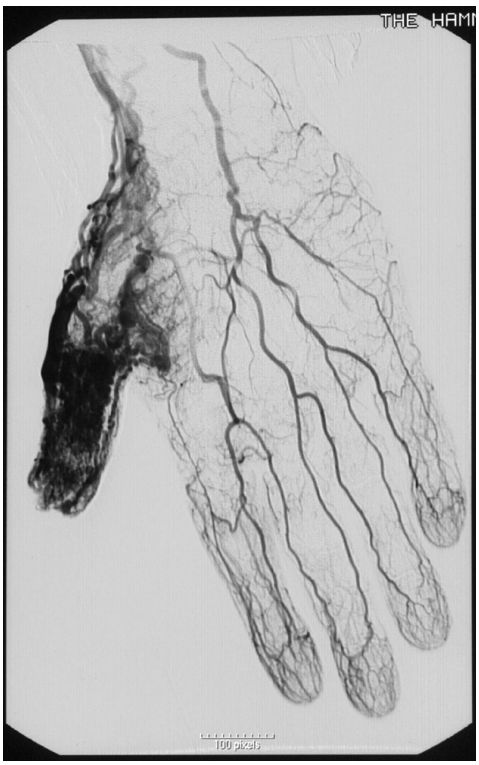

Figure 4. *Angiography under general anaesthetic demonstrated moderate hypertrophy of the radial artery and extensive abnormal increased vascularity throughout the right thumb with moderately rapid arteriovenous shunting into numerous dilated veins. Most of this abnormal vascularity was centred around and within the proximal phalanx but not confined to this area, with involvement also of all of the soft tissues of the digit extending to the pulp.*

Figure 5. *The component of this vascular malformation within the proximal phalanx was punctured percutaneously and partially and the intraosseous abnormality was embolised with 3% sodium tetradecyl sulphate. This final angiogram demonstrates some reduction in vascularity within the digit. It was decided that further embolisation thumb might compromise the vascular supply to the thumb.*

Discussion

Arteriovenous malformations can be congenital or acquired. Acquired lesions, more usually termed arteriovenous fistulas (AVFs) are associated with a history of significant penetrating trauma and which often occur during vascular interventional procedures. Congenital AVMs may not present for many years, or at all, depending on severity and precipitating events[2].

Arteriovenous malformations involving the digits are rare but, when present, may be complicated by ulceration[2]. This presentation (but not the underlying condition) is sometimes precipitated by trauma, which produces a wound which then fails to heal due to a combination of venous hypertension and pulp ischaemia secondary to the steal effect of the arteriovenous shunts[3]. Most individuals presenting with ulceration due to a digital AVM will either be aware of the presence of an underlying vascular malformation or will give a history of a pre-existing abnormality (e.g. overlying cutaneous birthmark; long-standing prominence of veins in the finger and hand) and the diagnosis in such cases is therefore straightforward. The diagnosis in the case presented here was especially difficult because of the absence of such a history and the very rapid development of ulceration without a clear history of significant trauma.

Management of digital AVMs is difficult and, in patients with few or no symptoms, it is often best to leave them untreated. When pain is severe or when ulceration occurs, however, treatment is mandatory. This may, in some cases, require digital amputation[4]. In some individuals, this can be avoided by therapeutic embolisation, which aims to occlude the AV communications within the malformation, whilst preserving normal digital arteries[5-7]. Complete obliteration of AV shunting is rarely achieved because of the intimate relationship between normal and abnormal vessels, but partial embolisation will often reduce venous hypertension within the digit sufficiently to allow skin healing. There are significant risks of compromising the vascularity of a digit with this technique, and this must be given careful consideration during open discussion with and informed consent from patients. However, the majority of patients derive significant benefit with relatively few complications when treated in specialist units[8].

Competing Interests - None declared.

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